In the claims:

Claim 1 (Previously Presented): A transgenic mouse wherein both alleles encoding Alpha Hemoglobin Stabilizing Protein (AHSP) have been disrupted via targeted insertion of a transgene wherein said mouse does not express a functional mouse Alpha Hemoglobin Stabilizing Protein (AHSP) protein and erythrocytes obtained from said mouse exhibit one or more characteristics selected from the group consisting of abnormal spiculated morphology, reduced lifespan, increased production of reactive oxygen species (ROS), and precipitated hemoglobin.

Claim 2 (Previously Presented): The transgenic mouse of claim 1, wherein said mouse transmits said transgene to its offspring.

Claim 3 (Previously Presented): The transgenic mouse of claim 1, wherein said transgene has been introduced into an ancestor of said mouse at an embryonic stage following microinjection of embryonic stem cells into a mouse blastocyst.

Claim 4 (Previously Presented): The transgenic mouse of claim 1, wherein said transgene has been introduced into an ancestor of said mouse at an embryonic stage following co-incubation of embryonic stem cells with a fertilized egg or morula.

Claim 5 (Previously Presented): A transgenic mouse wherein one allele of its endogenous Alpha Hemoglobin Stabilizing Protein (AHSP) gene has been disrupted via targeted insertion of a transgene wherein said mouse exhibits AHSP haploinsufficiency and has an elevated reticulocyte count.

Claim 6 (Previously Presented): The transgenic mouse of claim 5, wherein said mouse and transmits said transgene to its offspring.

Claim 7 (Previously Presented): The transgenic mouse of claim 5, wherein said transgene has been introduced into an ancestor of said mouse at an embryonic stage following microinjection of embryonic stem cells into a mouse blastocyst.

Claim 8 (Previously Presented): The transgenic mouse of claim 5, wherein said transgene has been introduced into an ancestor of said mouse at an embryonic stage following co-incubation of embryonic stem cells with a fertilized egg or morula.

Claim 9 (Original): A method for screening for therapeutic agents which affect Alpha Hemoglobin Stabilizing Protein (AHSP) activity, comprising:

a) administering a test compound to the transgenic mouse of claim 1;

b) assessing said mouse for an alteration in an Alpha Hemoglobin Stabilizing Protein (AHSP) activity.

Claim 10 (Original): The method of claim 9, wherein said activity is selected from the group consisting of α-hemoglobin binding, and α-hemoglobin synthesis.

Claim 11 (Previously Presented): A method for assessing the activity of a compound useful for the treatment and/or prevention of an AHSP related disorder, comprising:

a) providing at least one transgenic mouse as claimed in claim 1;

b) administering a test compound to said mouse; and

C) assessing the erthrocytes of said mouse for an alteration in one or more characteristics selected from the group consisting of abnormal spiculated morphology, reduced lifespan, increased production of reactive oxygen species (ROS), and precipitated hemoglobin, thereby identifying agents useful for treatment of an AHSP related disorder.

Claim 12 (Original): The method of claim 11, said method further comprising administration of said test compound to a control mouse to assess toxicity of said test compound.

Claim 13 (Cancelled)

Claim 14 (Withdrawn): A method of diagnosing an Alpha-Hemoglobin Stabilizing Protein (AHSP) related disorder in a test subject, wherein a sample from said test subject is analyzed by a method selected from the group consisting of:

a) a method employing a specific binding member capable of binding to a AHSP nucleic acid sequence, the specific binding member comprising nucleic acid hybridizable with the AHSP sequence; and

b) a method of determining the presence, in a sample from a test subject, of a polypeptide encoded by the AHSP nucleic acid and, if present, determining the expression level; and

c) a method wherein at least one antibody domain with specificity for an epitope selected from the group consisting of a native AHSP nucleic acid sequence epitope, or a polypeptide epitope, the specific binding member being labeled so that binding of the specific binding member to its binding partner is detectable; and

d) a method of PCR amplification involving one or more primers based on AHSP gene sequence to screen for an decrease in AHSP expression in a sample from a test subject; and

e) a method of determining the presence, in a sample from a test subject, of a polypeptide encoded by the AHSP nucleic acid and, if present, determining the presence of mutations of the AHSP nucleic acid.

Claim 15 (Withdrawn): The method of claim 14, wherein said disorder is selected from the group consisting of α-thalassemia, β-thalassemia, anemia, spongiform encephalopathy, prion disease, and Alzheimer's disease.

Claim 16 (Withdrawn): The method of claim 15, wherein said disorder is bovine spongiform encephalopathy.

Claim 17 (Withdrawn): A method of screening for compounds which modulate the activity of an AHSP polypeptide, the method comprising contacting at least one test compound with the AHSP polypeptide in a reaction medium, testing the activity of the treated AHSP polypeptide and comparing that activity with the activity of native, untreated AHSP polypeptide in a comparable reaction medium.

Claim 18 (Withdrawn): A compound identified by the method of claim 11 or 17, wherein said compound is a fragment of AHSP, or a small molecule which mimics AHSP activity.

Claim 19 (Withdrawn): A method of treating, or ameliorating symptoms of an AHSP related disorder comprising over expressing an AHSP encoding nucleic acid molecule in the cells or body fluid of a patient having said disorder.

Claim 20 (Withdrawn): The method of claim 19, wherein said disorder is selected from the group consisting of α-thalassemia, β-thalassemia, anemia, spongiform encephalopathy, prion disease, and Alzheimer's disease.

Claim 21 (Withdrawn): A method of treating, or ameliorating symptoms of an AHSP related disorder comprising administering the compound of claim 18 to the cells or body fluid of a patient having said disorder.

Claim 22 (Withdrawn): The method of claim 21, wherein said disorder is selected from the group consisting of α-thalassemia, β-thalassemia, anemia, spongiform encephalopathy, prion disease, and Alzheimer's disease.

Claim 23 (Withdrawn): A method for producing alpha hemoglobin stabilizing protein (AHSP)-specific antibodies, comprising:

a) immunizing the transgenic mouse of claim 1 with an immunogenic amount of AHSP or fragments thereof;

b) harvesting serum from said mouse; and

c) screening said serum for antibodies immunoreactive to AHSP.

Claim 24 (Withdrawn): An antibody preparation produced by the method of claim 23.

Claim 25 (Withdrawn): A method for producing alpha hemoglobin stabilizing protein (AHSP)-specific antibodies, comprising:

a) immunizing the transgenic mouse of claim 1 with an immunogenic amount of AHSP or fragments thereof;

b) harvesting the spleen of said mouse and fusing said spleen cells with a myeloma cell line containing a mutation to facilitate isolation of fused spleen/myeloma cells;

c) culturing said fused cells in media containing a selection agent wherein fused cells grow, and non-fused cells are killed;

d) screening said media from cells surviving in the presence of said selection agent, for the presence of antibodies immunoreactive to AHSP; and

e) optionally isolating said antibody.

Claim 26 (Withdrawn): A monoclonal antibody produced by the method of claim 25.

Claim 27 (Withdrawn): A kit comprising one or more molecules for detecting AHSP expression, said molecules being optionally detectably labeled, and said molecules being selected from the group consisting of nucleic acid molecules having sequences corresponding to a portion of an AHSP nucleic acid sequence for use in amplifying a nucleic acid comprising an AHSP nucleic acid sequence, and antibodies which specifically bind to a portion of the AHSP protein.

Claim 28 (Withdrawn): A transgenic mouse characterized by overexpression of an Alpha Hemoglobin Stabilizing Protein (AHSP) gene.

Claim 29 (Withdrawn): The transgenic mouse of claim 1, said mouse also having a heterozygous null mutation in beta major and minor globulin genes on a single chromosome, said mouse exhibiting at least one characteristic selected from the group consisting of low hematocrit level, and increased red cell distribution width (RDW).

Claim 30 (Withdrawn): A method for assessing the activity of compounds useful for the treatment and/or prevention of an AHSP related disorder, comprising:

a) providing mice as claimed in claim 29;

b) administering a test compound to the mice of step (a); and

C) assessing said mice for inhibition of said AHSP related disorder.

Claim 31 (Withdrawn): The method of claim 30, said method further comprising administration of said compound to control mice to assess toxicity of said test compound.

Claim 32 (Withdrawn): A compound identified by the method of claim 30.

Claim 33 (Withdrawn): A method of treating, or ameliorating symptoms of an AHSP related disorder comprising administering the compound of claim 32 to the cells or body fluid of a patient having said disorder.

Claim 34 (Withdrawn): The transgenic mouse of claim 1, said mouse also having a homozygous null mutation in at least one alpha globulin gene, said mouse exhibiting at least one characteristic selected from the group consisting of low hematocrit level, high reticulocyte count, decreased mean corpuscular volume (MCV), and increased red cell distribution width (RDW).

Claim 35 (Withdrawn): A method for assessing the activity of compounds useful for the treatment and/or prevention of an AHSP related disorder, comprising:

a) providing mice as claimed in claim 34;

b) administering a test compound to the mice of step (a); and

C) assessing said mice for inhibition of said AHSP related disorder.

Claim 36 (Withdrawn): The method of claim 35, said method further comprising administration of said compound to control mice to assess toxicity of said test compound.

Claim 37 (Withdrawn): A compound identified by the method of claim 35.

Claim 38 (Withdrawn): A method of treating, or ameliorating symptoms of an AHSP related disorder comprising administering the compound of claim 37 to the cells or body fluid of a patient having said disorder.

Claim 39 (Currently Amended): The method of claim 9, wherein said ~~agent~~ test compound alters one or more characteristics selected from the group consisting of abnormal spiculated morphology, reduced lifespan, increased production of reactive oxygen species (ROS), and precipitated hemoglobin in the erythrocytes of said mice.